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CASE REPORT

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Original Article

A Cross Sectional Study on Prevalence of JAK2V617F mutation in Primary mesenteric and Portal Venous Thrombosis at Tertiary care center, Jaipur

Nidhi Sharma*, Gaurav Kumar Gupta, Ishita Ishu Railja***, Sandhya Gulati****, Ankur Kumar*****, Peeyush Kumar*****

ABSTRACT

Background: Mesenteric venous thromboses (MVT) and portal venous thromboses (PVT) are uncommon diseases with excessive mortality rates.[1] JAK2V617F mutation finding is a significant diagnostic tool for patients of MPN. Through our study we aim to provide the importance of screening JAK2V617Fmutation in patients of PVT/MVT, which can be pivotal in preventing major bleeding episodes and their complications. Only in a few cases of MVT/PVT can be detected the clinical signs of an underlying the myeloproliferative neoplasm (MPN) or heritable thrombophilia.

Methodology: This descriptive analytical study included 100 patients with splanchnic venous thrombosis (MVT & PVT). All the blood samples (0.5ml EDTA peripheral blood) were received, processed and qualitative analysis for JAK2V617F mutation was performed in Advanced Hematology Lab, SMS Medical College, Jaipur. Those who were positive for JAK2V617F mutation were then advised bone marrow aspiration and biopsy.

Results: 100 radiologically confirmed cases of splanchnic venous thrombosis (mesenteric and/or portal) were taken. Out of these 100 cases, 36% had the JAK2V617F mutation. Polycythemia vera (PV) emerged as the leading (96%) Myeloproliferative neoplasm (MPN) in these 36% patients on examination of bone marrow biopsy and aspiration.

Conclusion: This study reveals the importance of JAK2V617F mutation testing in patients suffering from PVT and MVT. Almost one third of the patients had molecular evidence suggesting MPN (most commonly PV), highlighting the importance of this category of haematological neoplasm as a life-threatening intra-abdominal thrombosis cause.

Keywords: Portal venous thrombosis, JAK2V617F Mutation & Polycythemia vera.

INTRODUCTION

The JAK2V617F mutation, a visceral mutation in the JAK2 gene, is strongly related to myeloproliferative neoplasms (MPNs) and thrombosis, especially in splanchnic veins. This mutation results in increased cell proliferation and poses a risk to thrombosis through different mechanisms. The presence of JAK2V617F mutation in patients with primary mesenteric or portal venous thrombosis can signal an underlying MPN, even if it is not clinically evident.

In the western studies the generality of JAK2V617F mutation among Splanchnic venous thrombosis (SVT) patient is huge and extends from 7 to 59% but the relative frequency of this mutation amongst Indian SVT patients is heterogeneous.[2] No such data is available from north western India.

So the study aims to provide the importance of screening JAK2V617F mutation in patients of PVT/MVT, which can be pivotal in preventing major bleeding episodes and their complications.

MATERIALS AND METHODS

This was a cross-sectional study conducted at the Advanced Hematology Lab, Department of Pathology, SMS Medical College, Jaipur between March 2022 and April 2024. Elaborated Informed permission was taken from the patients or their attendants before the beginning of study. Ethics clearance was duly obtained from Institutional Ethics Committee. All consecutive eligible patients fulfilling following inclusion (with newly diagnosed mesenteric or portal venous thrombosis) and exclusion criteria (oral anti coagulation drugs and Hepatocellular Carcinoma) were included. 0.5ml of Peripheral blood was collected under aseptic conditions, after obtaining informed consent, into EDTA anticoagulant. TRUPCR® JAK2 KIT Version 2.0 was used for the extraction of DNA and Qualitative Detection of JAK2 V617F Mutation was done. Those who were positive for JAK2V617F mutation were then advised bone marrow aspiration and biopsy.

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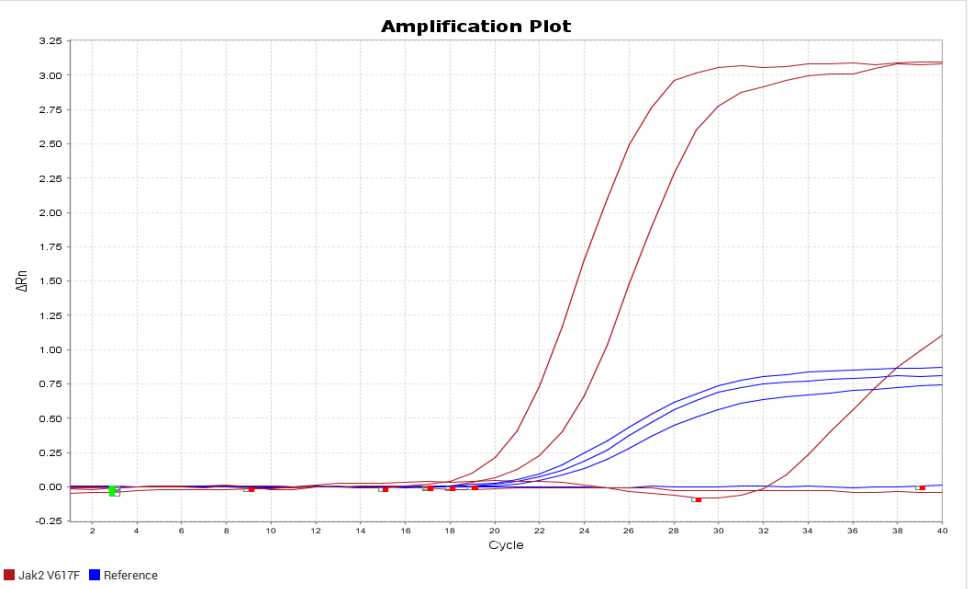
RESULTS

The study found a statistically significant difference in age distribution between JAK2V617F-positive and negative patients (p = 0.008). JAK2V617F- positive patients had a higher mean age of 45.72 years compared to 36.17 years for JAK2V617F- negative patients. This suggests that the mutation is more prevalent in older patients with primary mesenteric and portal venous thrombosis and the thrombosis in younger patients occurs due to other causes more commonly.

There was a significant association between organomegaly and JAK2V617F mutation status (p = 0.023). (TABLE-1) 42.9% of JAK2-positive patients exhibited organomegaly compared to 57.1% of JAK2-negative patients. This association suggests that the presence of organomegaly could be an indicator for JAK2V617F mutation testing in patients with splanchnic vein thrombosis.

FIG1: RT- PCR Cases of JAK2 Mutation with Control



#	Well	Sample Name	Target Name	Task	Cr	Quantity
5	A5	Jak2-121	Reference	UNKNOWN	22.852	
6	A6	Jak2-122	Jak2 V617F	UNKNOWN	32.332	
6	A6	Jak2-122	Reference	UNKNOWN	22.642	
7	A7	Jak2-123	Jak2 V617F	UNKNOWN	Undeter...	
7	A7	Jak2-123	Reference	UNKNOWN	Undeter...	
8	A8	Jak2-124	Jak2 V617F	UNKNOWN	32.353	
8	A8	Jak2-124	Reference	UNKNOWN	22.705	
9	A9	Jak2-125	Jak2 V617F	UNKNOWN	32.334	
9	A9	Jak2-125	Reference	UNKNOWN	22.799	
10	A10	Jak2-126	Jak2 V617F	UNKNOWN	32.131	
10	A10	Jak2-126	Reference	UNKNOWN	22.456	
11	A11	Jak2-127	Jak2 V617F	UNKNOWN	19.405	
11	A11	Jak2-127	Reference	UNKNOWN	22.146	
12	A12		Jak2 V617F	NTC	Undeter...	
12	A12		Reference	NTC	Undeter...	

Sample from JAK2-121 to JAK2-126 are negative for JAK2 mutation and JAK2-127 is positive for JAK2 mutation.

TABLE -1: Clinical and radiological findings in JAK2V617F mutation positive and negative patients.

			JAK2V617F		Total	p VALUE*
			POSITIVE	NEGATIVE		
ORGANOMEGALY	ABSENT	Number of patients	6	24	30	0.023
		%	20.00%	80.00%	100.00%	
	PRESENT	Number of patients	30	40	70	
		%	42.90%	57.10%	100.00%	
Total		Number of patients	36	64	100	
		%	36.00%	64.00%	100.00%	
PORTAL VEIN THROMBOSIS	NOT INVOLVED	Number of patients	5	22	27	0.027
		%	18.50%	81.50%	100.00%	
	INVOLVED	Number of patients	31	42	73	
		%	42.50%	57.50%	100.00%	
Total		Number of patients	36	64	100	
		%	36.00%	64. 00%	100.00%	

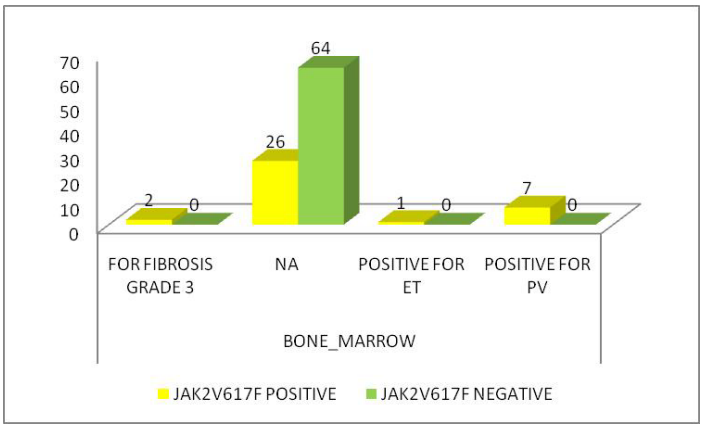


Figure 2: Bone marrow aspiration and biopsy finding in JAK2V617F mutation positive and negative patients.

Our study revealed significant associations between the JAK2V617F mutation and specific bone marrow findings. The study found a highly significant association between PV and JAK2V617F mutation ($p = 0.0001$). 90% of patients with PV were JAK2-positive, compared to only 30% of those without PV. This strong association reinforces the central role of the JAK2V617F mutation in the pathogenesis of PV and supports its use as a diagnostic criterion for this condition.

This strong association aligns with the findings of De Stefano V et al. (2007)[3] and underscores the close relationship between the JAK2V617F mutation and myeloproliferative neoplasms (MPNs).

DISCUSSION

Our study found a 36% prevalence of the mutation in splanchnic venous thrombosis patient population. This finding is compared with several previous studies to contextualize its significance**. Narendra Kumar et al. (2020)**[2]: reported a wide range (7-59%) of JAK2V617F mutation prevalence among splanchnic vein thrombosis (SVT) patients. The current study's finding of 36% falls within this range, suggesting consistency with other Indian studies. The wide range reported by Kumar et al. highlights the potential variability in prevalence even within the same country, which could be due to differences in patient populations, environmental factors, or genetic backgrounds across different regions of India. **Kiladjian et al. (2008)**[4]: Conducted a larger study involving 241 patients with both Budd-Chiari syndrome (BCS) and portal vein thrombosis (PVT). While they didn't report an overall prevalence for the JAK2V617F mutation, their study emphasized the diagnostic importance of this mutation in splanchnic vein thrombosis. The larger sample size in Kiladjian's study provides a broader context for the current findings, suggesting that the observed prevalence is consistent with what is seen in larger, diverse populations of splanchnic vein thrombosis patients. **X.Qi et al. (2011)**[5]: did a meta-analysis that reported varying prevalence rates of JAK2V617F mutation in different types of splanchnic vein thrombosis, with higher rates in BCS compared to PVT. The current study's prevalence of 36% falls within the range reported by X.Qi et al. for various types of splanchnic vein thrombosis. This consistency across different studies and populations underscores the robust association between the JAK2V617F mutation and splanchnic vein thrombosis. However, the variation in prevalence across different types of splanchnic vein thrombosis suggests that the mutation may play different roles in the pathogenesis of thrombosis in different vascular beds. These studies collectively provide context for the current research, highlighting the variability in JAK2V617F mutation prevalence across different populations and types of splanchnic vein thrombosis, while also reinforcing the importance of screening for this mutation with non invasive procedure in patients with these conditions.

CONCLUSION

This cross-sectional study provides significant insights into the prevalence and clinical implications of the JAK2V617F mutation in patients with primary mesenteric and portal venous thrombosis. The prevalence of 36% for the JAK2V617F mutation in our cohort emphasis its importance in the etiology of splanchnic vein thrombosis. Our findings reveal significant associations between the mutation and various clinical and hematological parameters, including older age, organomegaly, portal vein involvement, and elevated blood counts. The study also highlights the strong link between the JAK2V617F mutation and myeloproliferative neoplasms (MPNs), particularly polycythemia vera and essential thrombocythemia. Importantly, study observed an increased risk of bleeding events in JAK2-positive patients. These results emphasize the need for routine JAK2V617F mutation screening in patients with splanchnic vein thrombosis which would be helpful in diagnosing an underlying hematologic conditions.

CONFLICT OF INTEREST: There was no conflict of interest in the study undertaken.

ETHICS APPROVAL: Ethics Clearance was obtained from Institutional Ethics Committee – S.M.S. Medical College & Attached Hospitals, Jaipur.

INFORMED CONSENT: Written Informed Consent was obtained from all the patients or their attendants before the study.

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Original Article

Magnetic Resonance Imaging in Evaluation and Characterization of Sellar and Juxtasellar Lesions with Histopathological Correlation

Deepak Kumar Meena*, Anu Bhandari, Shubha Sharma***, Raghav Tiwari****, Mukesh Mittal****

ABSTRACT

Background: The wide spectrum of sellar and juxtasellar lesions often present with similar symptoms demonstrating profound neuroendocrine manifestations. Preoperative noninvasive diagnosis with magnetic resonance imaging (MRI) is essential for treatment planning. Thus this study intended to assess the accuracy of Magnetic Resonance Imaging in imaging of sellar and juxtasellar lesions taking histopathological findings as gold standard.

Methods: This cross sectional observational study was carried out during January 2023 to September 2023. Total 66 patients who were clinically suspected having sellar/juxtasellar lesions were included in the present study and underwent MRI examination. MRI was performed in GE Sigma Architect 64 Channel 3T MRI Machine.

Results: Out of 66 patients, 51.5% cases were males and 48.5 % cases were females. Most common pathology was adenoma (54.5%) followed by Craniopharyngioma (18.1%) and meningioma (10.6%). Histopathological correlation revealed MRI accuracy of 93.94%, 93.91%, 100%, 96.97%, 98.48% and 98.48% for the diagnosis of adenoma, craniopharyngioma, meningioma, pilocytic astrocytoma, apoplexy and epidermoid respectively.

Conclusion: The present study revealed a strong correlation between MRI and histopathological diagnosis for sellar and juxtasellar leisons. MRI is the modality of choice for characterizing sellar and suprasellar lesions.

Keywords: MRI, Magnetic resonance imaging, Macroadenoma, Craniopharyngioma Meningioma.

INTRODUCTION

The sella turcica and adjoining area is a small but complex component of the central nervous system, containing many vital structures. The wide spectrum of sellar and juxtasellar lesions often present with similar symptoms demonstrating profound neuroendocrine manifestations. Early diagnosis and accurate characterization are able to provide significant clinical benefit. While definitive diagnoses usually await histologic correlation, a systematic approach and knowledge of the key anatomy and imaging features can help in narrowing down the differential diagnosis and sometimes to reach a specific one. [1]

At least 30 different lesions occur in or around the pituitary gland, arising from either the pituitary gland itself or the structures that surround it. [2] Pituitary tumors account for up to 15% of all intracranial masses, and at least 75-80% of all sellar/juxtasellar masses are due to one of the "Big Five": Adenoma (most common), meningioma, aneurysm, craniopharyngioma, and astrocytoma. Clinically active pituitary adenomas occur at a prevalence of 1:1064 to 1:1288 to the general population. Other sellar lesions include nonneoplastic cystic lesions, germ cell tumors, gliomas, lymphomas, meningiomas, metastatic tumors, vascular lesions, granulomatous and inflammatory lesions, and infections including bacterial abscesses as well as pituitary hyperplasia.[3]

Magnetic resonance imaging has virtually supplanted other imaging techniques such as CT as the modality of choice for evaluation of the sellar and juxtasellar regions Its major advantages are its superior soft tissue contrast and capacity for multiplanar imaging.[4] Dynamic MR Imaging plays a very important role in the evaluation of pituitary adenomas, particularly in accurate delineation of those microadenomas with no contour abnormality, to assess invasion by macroadenoma and in post-operative evaluation in cases of residual / recurrent lesions.

In India, the prevalence in Rajasthan was 7.6%. Unfortunately, no nationwide prevalence and incidence data is available from Indian cohort .[5] However, there is a dire need for country-specific research to formulate patient treatment and management guidelines. A multi-disciplinary approach involving endocrinologist, neurosurgeon, Radiologist, histopathologist etc. would be very helpful.

Thus this study is intended to assess the accuracy of Magnetic Resonance Imaging in imaging of sellar and juxtasellar lesions taking histopathological findings as gold standard.

METHODS

This cross sectional observational study was carried out during January 2023 to September 2023 in the department of Radiodiagnosis, S.M.S. Medical College and Hospitals, Jaipur. Total 66 patients who were clinically suspected having sellar/juxtasellar lesions were

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included in the present study and underwent MRI examination. Claustrophobic patients and patients with general contraindications to MRI such as pacemakers, metallic fixations, etc. were excluded from the study. The procedure was communicated to the patients and the informed consent was taken in written format. A brief history and examination was done at the time of MRI scan. MRI was performed in GE Sigma Architect 64 Channel 3T MRI Machine. Different MRI features in terms of T1W and T2W signal characteristics (iso/hypo/hyperintensity), consistency, enhancement, and involvement of adjacent structures were noted in the structured format. The consistency of different lesions was categorized into solid, cystic, Solid - Cystic and Cystic with Mural nodule. The diagnostic accuracy of MRI in assessing sellar/juxtasellar lesions was compared with histopathological finding taken as gold standard.

RESULTS

The findings of the Sixty-six patients were compiled and analyzed. Out of 66 patients, 51.5 % cases were males and 48.5 % cases were females. Maximum number of cases were in the age group of 31-40 years (28.8%), followed by patients in the 51-60 years age group (19.6%). Over 50 per cent cases (~57.5 %) were in 11 to 40 years age group. Youngest patient was 4 years old and oldest patient was 71 years old,

Table- 1 shows the clinical feature of the patients with sellar lesion.

Most common symptoms observed were headache (62%) and diminution of vision (51.5 %). These both were together observed in 37.8 per cent cases.

Table 1 – Distribution of respondent according to clinical feature:

Clinical Feature	No. of Cases	Percentage
Headache	43	62
Diminution of vision	34	51.5
Combined (headache & diminution of vision)	25	37.8
Acromegaly	4	6
Cushing syndrome	4	6
Infertility/erectile dysfunction/decreased libido/Menstrual irregularities	4	6
Others – vomiting/ limb weakness/ seizures/ polyurea/polydipsia	8	12

Among all the Sellar, suprasellar and para-sellar lesions, 50 per cent cases showed elevated pituitary hormones level and 3.1 per cent decreased level whereas 46.9 per cent cases showed normal level.

Majority of pathologies in sellar and juxtasellar regions were neoplastic (~89%).

Most common pathology was adenoma (54.5 %) followed by Craniopharyngioma (18.1%) and meningioma (10.6 %). Except one pathology (two microadenomas were seen in a single patient), all other pathologies had single lesion. Over 85 per cent pathologies (~87.5 %) had well defined margins. Further, approximately 90% pathologies had both suprasellar and sellar components. [Table 2]

Table 2 – Distribution Of Pathologies in Sellar and Juxta-Sellar Regions

Pathology	No. of cases	Percentage
Adenoma	36	54.5
Craniopharyngioma	12	18.1
Meningioma	7	10.6
Pilocytic Astrocytoma	3	4.5
Pituitary Apoplexy	2	3
Epidermoid Cyst	2	3
Rathke Cleft Cyst	1	1.5
Tuberculoma	1	1.5
Lymphocytic Hypophysitis	1	1.5
Enchondroma	1	1.5
Total	66	100

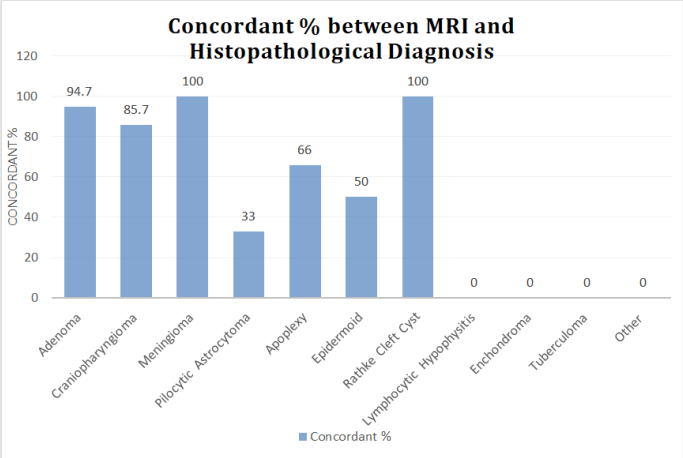
Majority of pituitary adenomas were macroadenomas (88.8%). Among macroadenomas, ~37.5 % were giant macroadenomas (> larger than 4 cm in at least one dimension). The second most common tumor was craniopharyngioma, most of the cases were children (75 %) and among them majority of were in 5 to 15 years age group (50 %). Youngest and oldest patients were of 4 and 51 years of ages respectively. No case was seen in 20 to 30 years and above 60 years age groups. All of the meningioma cases were in adult age group, youngest patient was of 37 years age and oldest 61 years.

All of the adenoma showed contrast enhancement, ~58 % heterogeneously enhancing and ~42 % homogeneously enhancing. Similarly, all craniopharyngioma and meningioma showed contrast enhancement, where most of craniopharyngioma manifested as rim/nodular enhancement (~83%) and 71% meningioma showed homogeneous enhancement followed by heterogeneous and rim enhancement.

Out of 66 Sellar/Juxta sellar lesions, ~56 % were isointense on T1W images followed by ~28 % hypointense and rest ~16 % hyperintense. Majority of adenoma (~66%) and meningioma (~85%) were isointense and Craniopharyngioma (~41%) were hyperintense on T1W images. Whereas, Most of adenoma (~66%) and Craniopharyngioma (~91%) were hyperintense and ~71 % meningioma was isointense. None of the Adenoma, Craniopharyngioma and meningioma were hypo, iso and hyperintense on T2W respectively. Most of the adenoma cases (75%) and meningioma cases (85.71%) were solid in consistency, whereas 41.67% of craniopharyngioma cases were mixed solid-cystic in consistency. [Table 3]

In our study, high concordance percentage was observed for Meningioma (100%), Adenoma (94.7%), Craniopharyngioma (85.7%) and Rathke Cleft Cyst (100%), moderate for Apoplexy (66%) and Epidermoid (50%) and low for Pilocytic Astrocytoma (33%), lymphocytic Hypophysitis, Tuberculoma & Enchondroma. [figure 1]

Figure 1 – Concordant Percentage Between MRI Diagnosis & Histopathology



Out of total 38 cases of MRI diagnosis of Adenoma, 3 turned out to be another pathologies – Tuberculoma, Enchondroma and Lymphocytic Hypophysitis. Further one case of MRI diagnosis of nasopharyngeal SOL came out as Adenoma. Whereas, out of total 14 cases of MRI diagnosis of Craniopharyngioma, 3 turned out to be another pathology- two Pilocytic Astrocytoma and one Epidermoid Cyst. Further one case of MRI diagnosis of Pituitary Apoplexy came out as Craniopharyngioma. All of the Meningioma cases were diagnosed correctly on MRI and there was no false negative case leading to 100 % - sensitivity, specificity, PPV, NPV and accuracy parameters. There was only one case of Rathke Cleft cyst which was diagnosed correctly. None of the Lymphocytic Hypophysitis, Tuberculoma or Enchondroma cases were diagnosed correctly on imaging. [Table 3]

Table 3 - Accuracy Parameters of MRI in Diagnosing Major Sellar/Juxta-Sellar Pathologies Taking Histopathology as Gold Standard

Pathology	Sensitivity	Specificity	Positive Predictive Value	Negative Predictive Value	Accuracy
Adenoma	97.22	90	92.11	96.43	93.94
Craniopharyn-gioma	91.67	94.44	78.57	98.08	93.91
Meningioma	100	100	100	100	100
Pilocytic Astrocytoma	33.33	100	100	96.92	96.97
Apoplexy	100	98.44	66.67	100	98.48
Epidermoid	50	100	100	98.46	98.48

All values are in %.

DISCUSSION

Diagnosing Sellar and Juxta-Sellar pathologies can be quite complex. The Sellar region, located in the central skull base, houses the pituitary gland and is surrounded by critical structures such as the optic chiasm, cavernous sinuses, and carotid arteries. This intricate anatomy makes imaging and interpretation challenging.

Clinical profile, lesion morphology, signal intensity on various sequences, and contrast enhancement pattern are taken into consideration when characterizing masses with MRI; however, even if the data are evaluated together, there can still be difficulties in the differentiation of various Sellar and Juxta-Sellar pathologies. The major aim of this study was to describe MR imaging characteristics of the lesions of the Sellar and Juxta-Sellar region; to correlate the MRI findings with histopathological findings and to assess the diagnostic accuracy of MRI in characterization of these lesions.

The patients belonged to all age groups ranging from 4 to 71 years in our study. However, Maximum number of cases were in the age group of 31-40 years (28.8%), followed by patients in the 51-60year age group (19.6%), 21-30year age group (15.1 %) and 11-20year age group (13.6%).

Our results are consistent with the study conducted by Dogar T et al[6], Banna et[7] al, Goyani BR et al[8], Prerit J et al[9], Batra V et al[10], Karthikeyan V et al[11], Omprakash et al[12], Pratisruti Hui et al[13] and Kaushal B et [14], who also encountered maximum number of patients in these age groups.There was slight male preponderance (male-female ratio - 1.06:1) in our study which was similar to Karthikeyan V et al[11] and Ompraksh et al[12] experiences; in contrast to Banna et al[7], Kaushal et al[14], Batra V et al[10], Pratisruti Hui et al[13], Suman et al[15] and Prerit J et al[9] who reported a female preponderance.

Headache (62%), followed by visual disturbances (51.5 %) and combined (headache and visual disturbance) (31.5%) were the most common symptoms in our study, and other complaints were hormonal imbalance leading to amenorrhea, galactorrhea, infertility, erectile dysfunction, Cushing syndrome and acromegaly and mass effect causing seizures, hemiparesis, and vomiting, these findings are similar to the study done by Karthikeyan V et al[11], Pratisruti Hui et al[13] and Batra et al[10]. This particular involvement of symptoms is explained by the fact that most of the Sellar lesion have suprasellar extensions which causes mass effect leading to raised intracranial pressure so headache and compression and elevation of optic chiasm so visual disturbances.

Our study observed that pituitary adenomas, craniopharyngioma, and meningioma were the major constituents for the study consisting of 54.5%, 18.1%, and 10.6% of the cases, respectively, followed by pilocytic astrocytoma (4.5%), epidermoid (3%), and apoplexy (3%). There was one case of each lymphocytic hypophysitis, tuberculoma,rathke cleft cyst and enchondroma.

These findings are similar to what was observed by Johnsen et al[3], Batra V et al[10], Omprakash et al[12], Pratisruti

Hui et al[13], Karthikeyan V et al[11], Kaushal B et al[14], Hemalatha et al[16], Batra V et al[10] and Prerit J et al[9] who all observed that the most common lesion in Sellar and Juxta-Sellar region was adenoma followed by craniopharyngioma and meningioma. Majority of pathologies in sellar and juxtasellar regions were neoplastic (~89%) in our study consistent with other above-mentioned studies.

Pituitary Adenoma

In the present study, there were 36 pituitary adenoma cases which constituted about 54.5% of total pathologies in Sellar and Juxta-Sellar regions. Maximum number of cases were in the age group of 31-40 years (41.6 %). Out of 36 adenomas 32 were macroadenomas and 4 were microadenomas by size criterion. The proportion of macro to microadenoma was 8:1. Johnsen et al found the total proportion of macro to microadenomas 2.5:1 while Kaushal B et al observed it to be 10.5:1. Though literature says that microadenomas are more common than macroadenoma in general population.

High signal intensity on T1W images may be seen due to presence of hemorrhage in macroadenoma. We found that 24 cases (66 %) of macroadenomas were isointense to grey matter, 8 (22%) were hypointense and 4 (12%) were hyperintense on T1W images, and on T2W images 24 cases (66.6%) of macroadenomas were hyperintense to grey matter, 12 (33.3%) were isointense and none was hypointense.

Our findings of enhancement pattern of macroadenoma were similar to findings observed by Johensen et al[3] and Kaushal B et al[14]. The presence of cystic/necrotic/ haemorrhagic areas were reasons for heterogeneous enhancement. In our study in about 37.5% of the cases, internal carotid artery (ICA) encasement is noted which is supported by Johnsen et al[3] and Pratisruti Hui et al[13] where it was 43% and 42% respectively.

Our diagnostic accuracy parameters (sensitivity, specificity, PPV, NPV & accuracy - ~97%, 90%, 92%, 96% & 94%) are similar to what were observed by Kaushal B et al[14], Batra V et al[10].

Craniopharyngioma

Majority of the craniopharyngioma cases have peak age of occurrence at 5–15 years, followed by a small peak in 45–60 years' age group, and have equal sex distribution supporting our findings. In the present study having total 12 Craniopharyngioma cases, most of were children (75 %) and among them majority of were in 5 to 15 years age group (50%). Male-female ratio was 1: 1, therefore no gender predilection was noted in our study. On T1W images 5 cases (41%) of macroadenomas were hyperintense to grey matter, 4 (34%) were hypointense and 3 (25%) were isointense. Whereas, on T2W images all except one hypointense case were hyperintense; similar to what was shown by, Batra V et al[10] and Suman et al[15]. Our data were correlated with the study by Pratisruti Hui et al[13] where they also didn't find any completely solid Craniopharyngioma case and most common consistency was solid-cystic. All of the cases showed enhancement, most common pattern was peripheral rim/nodular enhancement followed by heterogeneous enhancement. These observations are consistent with the findings observed by Kaushal B et al[14].

Out of total 14 cases of MRI diagnosis of Craniopharyngioma, 3 turned out to be another pathology-two Pilocytic Astrocytoma and one Epidermoid Cyst. Further one case of MRI diagnosis of Pituitary Apoplexy came out as Craniopharyngioma. Our diagnostic accuracy parameters (sensitivity, specificity, PPV, NPV & accuracy - ~92%, 94%,78%,98%&94%) are similar to what were observed by Kaushal B et al[14], Batra V et al[10].

Meningioma

Meningiomas in Sellar region may arise from tuberculum sellae, diaphragmatic sellae, clinoid processes, optic nerve sheaths and wings of sphenoid. There were seven histopathologically proven meningiomas in our study. male-female ratio was 0.4:1 which was supported by Johnson et al[3], Kaushal B et al[14].

Meningiomas typically show iso-intensity on T1- and T2-weighted sequences with characteristic homogeneous enhancement. In our study, all showed iso-intensity on T1W; on T2W 71 % were isointense and rest hyperintense. All of meningioma cases showed enhancement; 71 per cent as homogeneous enhancement and rest as heterogeneous enhancement and all were solid in consistency Similar findings were observed in Batra V et al[10], Hemalatha et al[16] and Suman et al[15].

All of our meningioma cases were diagnosed correctly on MRI and had 100% concordance rate with histopathology so our diagnostic accuracy parameters stand as (sensitivity, specificity, PPV, NPV & accuracy – 100% for all). Similar results were observed by Pratisruti Hui et al[13] and Suman et al[15]. Hemalatha et al[16] recorded 90% sensitivity and 100% NPV in diagnosis of meningioma in her study.

CONCLUSION

MR imaging demonstrated distinct characteristics that enabled effective differentiation among various sellar and juxtasellar pathologies. The diagnostic accuracy of MRI for pituitary adenomas and sellar region tumors was strongly validated by histopathological correlation, showing high sensitivity and specificity. Hence, MRI can be considered as the modality of choice for diagnosing sellar and suprasellar, lesions with high accuracy, sensitivity, and specificity.

Abbreviations

CT: Computed Tomography
ICA: Internal Carotid Artery
MRI: Magnetic Resonance Imaging
NPV: Negative Predictive Values
PPV: Positive Predictive Values

Declaration of Acknowledgment

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Availability of data and materials

Data will be available by emailing deepak2011meena@gmail.com

Authors' Contributions

- Author1: Literature search, conceptualization, methodology, data acquisition, manuscript preparation and editing.

- Author2: Conceptualization, supervision, validation, review and editing.

- Author3*: Writing, reviewing and editing.

- Author4: Data collection.

- Author5: Data analysis and interpretation.

All authors have read and agreed to the published version of the manuscript. All authors have read the final manuscript.

Ethics approval and consent to participate

The research adhered to the ethical principles outlined in the Declaration of Helsinki (2013). Approval for the study protocol was granted by the S.M.S. Medical College and Attached Hospitals, Jaipur Ethics Committee, with a decision number of 421 in 2023. Electronic signatures were obtained as informed consent from all individual participants involved in the study. Every procedure conducted during the study complied with the standards of S.M.S. Medical College and Attached Hospitals, Jaipur Ethics Committee and aligned with the principles set forth in the 1964 Helsinki Declaration, along with its subsequent amendments.

Competing Interest

The authors declare that they have no competing interests.

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Original Article

Outcome Analysis of Patients with Extra Pulmonary Drug-Resistant Tuberculosis at The Nashik Nodal DRTB Centre

Komal Shah*, Bhavik Shah**

ABSTRACT

Background: As per Global tuberculosis report 2024 (1), India contributes 26% of Global TB incidence and 27% of MDR TB globally. Mortality due to TB (non HIV) was reported more than 3 lakhs (29% of global TB deaths). (2) Limited data exists on the prevalence and drug resistance patterns in extrapulmonary tuberculosis (EPTB). This study aims to assess the drug resistance patterns of Mycobacterium tuberculosis in EPTB cases and evaluate treatment outcomes.

Materials and Methods: This retrospective study was conducted to analyse patients diagnosed with extrapulmonary multidrug-resistant tuberculosis between April 2019 and March 2020. The objective was to assess the clinical profile and treatment response in these cases.

Results: A total of 54 patients were diagnosed with extrapulmonary drug-resistant TB within 1 year (April 2019 to March 2020). Of these, 66.6% were female, and 33.3% were male, with the most affected age group being 21-29 years (26.4%). Lymph node involvement was the most common manifestation followed by bones and joint involvement. Surprisingly, more than 70% patients were primary extrapulmonary DRTB patients with only 3 patients having history of TB contact. Rifampicin resistance was found in 74% of cases and 11% patients diagnosed as pre-XDR. Treatment yielded favorable outcomes in most patients with less than 10% mortality rate.

Conclusion: Among 54 EP-DRTB patients, 70% were primary cases—an unusual and notable finding. Despite this, treatment outcomes were positive, reflecting effective management at the Nodal DRTB Centre, Nashik.

INTRODUCTION

The WHO Global TB Report 2024 states that extrapulmonary TB accounted for 17% of the 7.5 million global TB cases in 2022, with a growing concern over drug-resistant forms. (1) India saw a 17.7% decline in TB incidence since 2015, yet continues to bear the highest burden globally. (2) Drug-resistant extrapulmonary TB poses a unique challenge in diagnosis and management, requiring intensified surveillance and tailored treatment strategies. This study aims to evaluate the clinical presentation and treatment outcomes of EP DR-TB under programmatic management at a nodal DRTB centre, Nashik.

MATERIALS AND METHODS

This retrospective study was conducted at the Nodal Drug-Resistant Tuberculosis (DR-TB) Centre, Department of Respiratory Medicine, Dr. Vasantrao Pawar Medical College, Nashik. The study included patients diagnosed with extrapulmonary multidrug-resistant (MDR) and extensively drug-resistant (XDR) tuberculosis between April 2019 and March 2020. Data were retrospectively extracted from patient records in accordance with the programmatic DR-TB management guidelines.

Inclusion Criteria:

Patients diagnosed with extrapulmonary MDR/XDR-TB and admitted between April 2019 and March 2020.

Exclusion Criteria:

Patients with incomplete medical records.

The primary aim was to determine the proportion of extrapulmonary drug-resistant TB cases and analyze their clinical profiles and treatment outcomes. Data were analyzed using appropriate statistical methods.

RESULTS

1) Demographic Variables:

From April 2019 to March 2020, 54 patients were diagnosed with extrapulmonary drug-resistant TB. Among them, 18 were male (33.33%) and 36 were female (66.66%). The most affected age group was 21-29 years (46.29%), followed by 10-20 years (25.92%). The least affected age group was 60-69 years (1.85%).

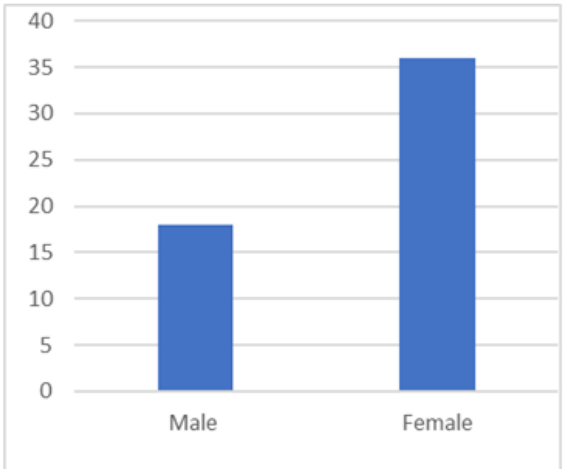

Fig.1 : Sex Distribution

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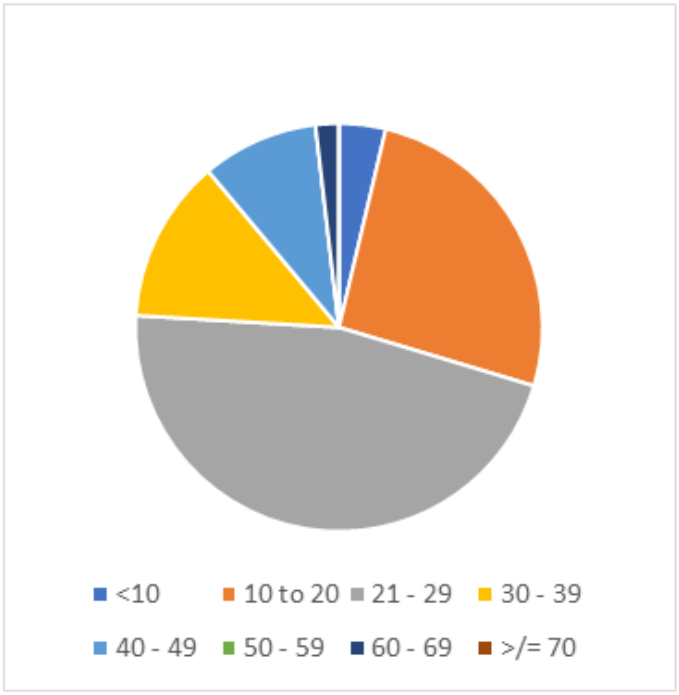

Fig.2 : Age group distribution

2) Organ Involvement:

Lymph node involvement was the most common site, occurring in 38 cases (70.37%), followed by bone involvement (13%). Resistance in breast abscesses and the central nervous system was also observed.

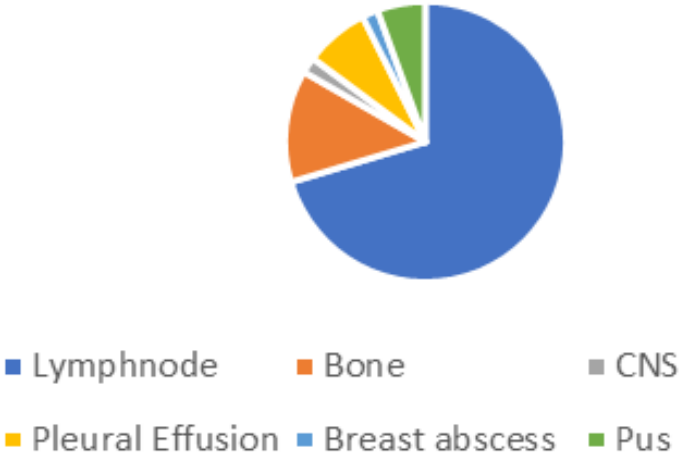

Fig.3 : Organ Involvement

3) History of Tuberculosis:

Eight patients had a history of pulmonary TB, and seven had a history of extrapulmonary TB. Three patients had known contact with DS/DR-TB cases.

4) Drug Resistance Pattern:

Among the study population, 74% were resistant to rifampicin, while 11.11% were classified as pre-XDR with fluoroquinolone resistance. Resistance to fluoroquinolones and isoniazid was equally observed.

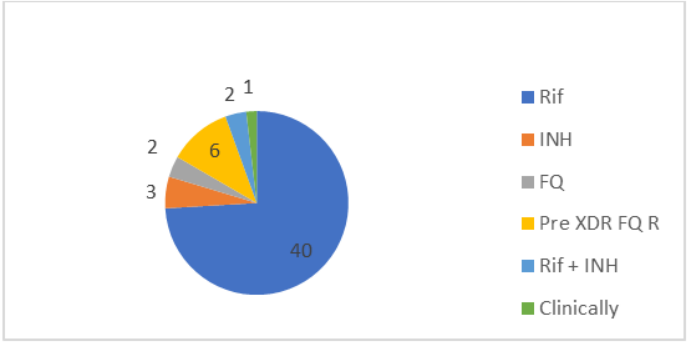

Fig.4 : Drug Resistance Pattern

5) Treatment Outcomes:

Treatment completion: 42 patients (77.77%)

Mortality during treatment: 5 patients (9.25%)

Defaulters: 5 patients (9.25%)

Lost to follow-up: 1 patient (1.85%)

Transferred out: 1 patient (1.85%)

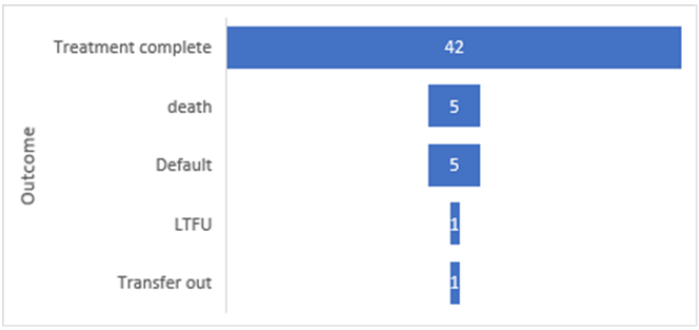

Fig.4 : Outcome

DISCUSSION

The proportion of extrapulmonary DR-TB cases in our study was 8.85%, higher than the 4.4% reported in previous studies. (3) The expansion of universal drug susceptibility testing has contributed to improved detection of DR-TB cases and resistance patterns. We also noticed higher rates of primary extrapulmonary DRTB cases than in previous studies. (5,6)

The majority of EP DR-TB cases were seen in younger patients (72%), with a higher prevalence in females (66%), aligning with findings from other studies. Lymph node involvement was the most common site (70.37%) same as other studies (7), followed by bone TB, differing from previous studies by the National Institute for Research in Tuberculosis (NIRT). (8)

Resistance to rifampicin was the most common, followed by fluoroquinolone resistance, differing from studies that reported higher pre-XDR fluoroquinolone resistance. HIV co-infection was found in four cases (7.40%), with one patient initiating treatment during pregnancy.

The treatment success rate was 77.77%, with 9.25% mortality and defaulter rates.

The retrospective nature of this study limited the ability to assess reasons for treatment default and mortality causes.

CONCLUSION

This study highlights the significant burden of extrapulmonary DR-TB, particularly in younger, female patients even in absence of prior history of TB or significant TB contact (i.e primary extrapulmonary DRTB). While most patients responded well to treatment, the high rates of rifampicin resistance and pre-XDR-TB require enhanced surveillance and individualized treatment strategies. Continued efforts to reduce treatment default and mortality are essential to improving outcomes in this population.

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CASE REPORT

Giant Chorioangioma of Placenta

Mukesh Mittal*, Pankaj Kumar Nitharwal, Dinesh Kumar Sahu***, Aruna Yadav*****

ABSTRACT

Chorioangiomas are rare benign tumors of the placenta, characterized by atrio- venous shunting within placenta leading to foetal anemia, cardiomegaly and hydrops. Maternal complications can be polyhydramnios, antepartum haemorrhage and Mirror syndrome. They are usually seen after 20 weeks, and most of them remain small and are asymptomatic. Large ones (> 4 cm) are termed as Giant Chorioangioma and can lead to foetal and maternal complications.

The proximity of the chorioangioma to the placental cord insertion site and its size determines its prognosis. Prenatal therapy in the form of interventions like direct injections of various chemicals and laser coagulation of the feeding vessels of the tumor can be done to reduce the foetal and maternal complications of giant chorioangiomas. Conservative management is done in cases of small chorioangiomas. We report a case of 27 year old female presented to us with history of 8 month amenorrhea for routine antenatal ultrasound examination and diagnosed as a case of Giant Chorioangioma. Early diagnosis was made followed by close surveillance with PSV of Doppler of MCA was done for foetal wellbeing and patient was managed conservatively with counseling of patient for any foetal and maternal complications.

INTRODUCTION

Placental chorioangioma also known as placental haemangioma is a non-trophoblastic benign tumor of the placenta that is characteristically vascular and originates from primitive chorionic tissue. It is rare and occurs in less than 1% of all pregnancies while incidence of Giant Chorioangioma is one in 9000 to one in 50000 pregnancies. Three types are recognized as Angiomatoid: characterized by numerous blood vessels, Cellular: with poor vascularisation and Degenerative: with degenerative changes.[1]

Placental chorioangiomas tend to occur on the foetal side of the placenta and close to cord insertion. Small sized less than 4 cm chorioangiomas are nonsignificant. Giant Chorioangiomas are larger than 4 cm in size and increases the risk of both foetal and maternal complications. Maternal complications can be preterm labour, placental abruption, polyhydramnios, and foetal complications can be foetal hydrops, intrauterine growth restriction, foetal anaemia, cardiac failure, and even foetal demise can occur in Giant chorioangiomas. Perinatal mortality has been reported in about 30-40% cases in Giant Chorioangioma. Therefore, prenatal diagnosis is vital to follow up these pregnancies diligently.[2]

CASE REPORT

A 27 year old G2P1, women with a previous full term normal vaginal delivery came for routine antenatal ultrasound examination at 8 month of amenorrhea. Her previous antenatal scan in first trimester and other blood investigations were normal. There was no significant family history of any placental or foetal anomaly. On ultrasound a well defined homogenous hypoechoic mass lesion of 11 x 8 cm size with few internal cystic areas was seen arising from foetal surface of placenta and projecting into the amniotic cavity showing mild vascularity with low resistant pulsatile flow on colour doppler. Other findings were foetal cardiomegaly with dilated right atria and backflow of blood in right atrium during ventricular contraction with regurgitation velocity more than 80cm suggestive of tricuspid regurgitation. Foetal complications like mild foetal ascites were noted. There were no maternal complications. This case was diagnosed as cellular type of Giant Chorioangioma.

DISCUSSION

Placental chorioangioma is the most common benign tumor of the placenta with reported incidence of approximately 1% of all pregnancies while incidence of

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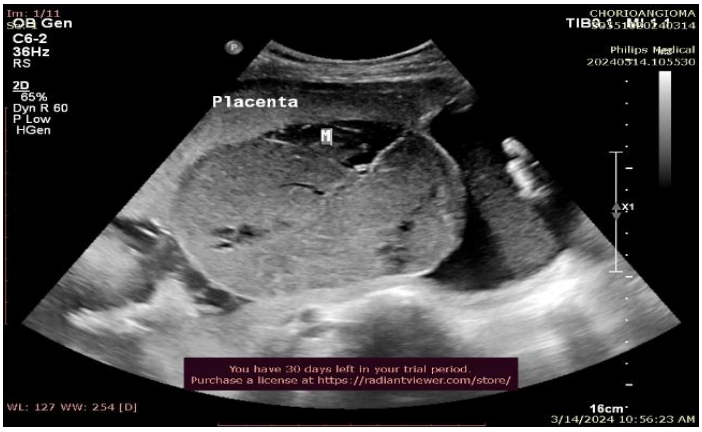
Giant Chorioangioma is one in 9000 to one in 50000 pregnancies. Most Chorioangiomas are small and asymptomatic. Large tumors more than 45 cm are associated with maternal and foetal complications. The pathophysioloy of the complications is not fully understood. The main hypotheses are: chronic arterio-venous malformation with a development of a high output foetal cardiac failure and the sequestration of red blood cells and platelets within the tumor. Antenatally it is diagnosed by ultrasound as a hypo or hyperechoic rounded mass with anechoic spaces which protrude into the amniotic cavity[3]. Most of them are located near the umbilical cord insertion. Color Doppler displays presence of vascular channels in the tumor contiguous with the foetal circulation. The most common 14-28% fetal complications are polyhydramnios, others are preterm delivery, non-immune hydrops, fetal cardiomegaly, fetal heart failure, fetal anemia, thrombocytopenia, fetal growth retardation and intrauterine fetal demise[4]. The maternal complications include pre-eclampsia and maternal mirror syndrome (maternal edema associated to foetal hydrops). The chorioangioma is usually managed conservatively[5]. Patients are monitored by ultrasound, where foetal growth, Doppler examination, signs of hydrops and tumor size are assessed. The most common invasive therapeutic procedures are amniocentesis and intrauterine fetal blood transfusion. Other invasive interventions to block AV shunting include alcohol injection, micro coil embolisation and endoscopic laser coagulation. These techniques are more successful when tumor is located away from umbilical cord site and its circulation is not directly depended on the umbilical cord[6].

CONCLUSION

Placental chorioangioma is certainly the most common benign tumor of placenta. Early diagnosis of placental tumor, its placental location, its composition and size of tumor with vascularity helps in reaching the final diagnosis of Giant chorioangioma. Regular ultrasound monitoring of foetal and maternal complications of Giant Chorioangiomas helps in timely intervention and thereby improving the prognosis.



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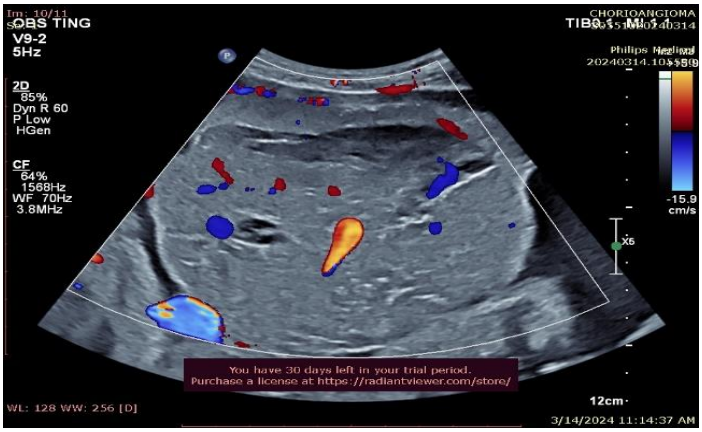

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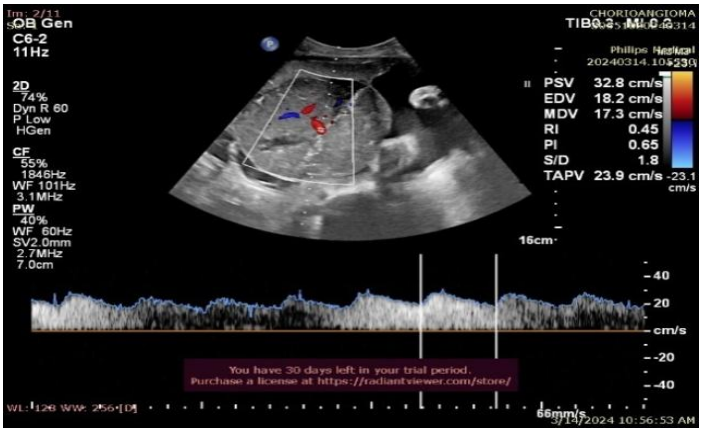

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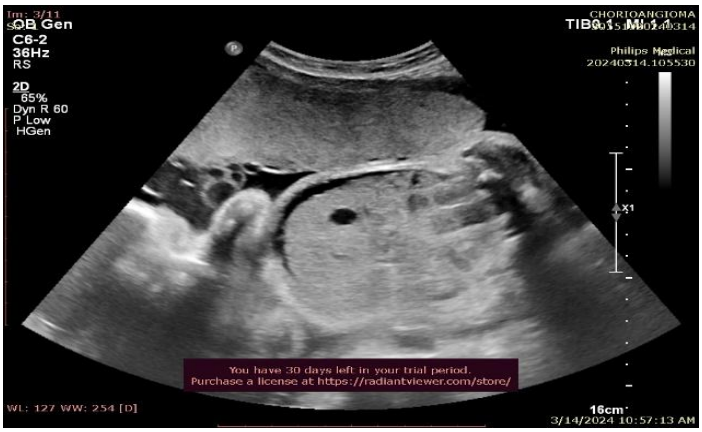

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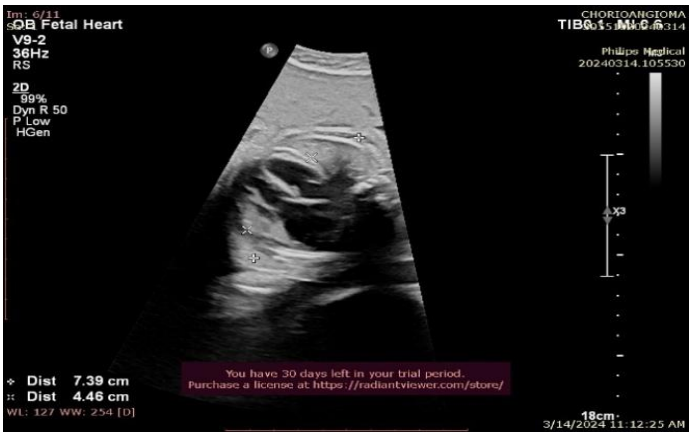

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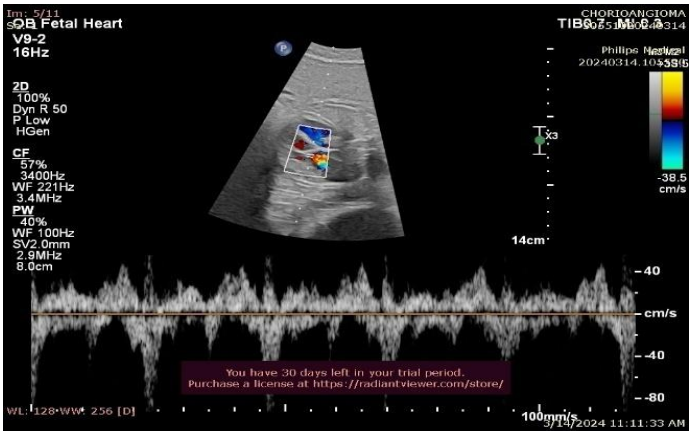

F

Fig 1 On USG a well defined homogenous hypoechoic sub chorionic mass lesion on foetal side of placenta with few internal cystic areas (A), showing vascularity (B) and low resistant pulsatile flow on colour doppler represent enlarged vascular channels (C) with mild foetal ascites (D), increased cardiothoracic ratio represent cardiomegaly (E), backflow of blood in right atrium during ventricular contraction with regurgitation velocity more than 80cm suggestive of tricuspid regurgitation.

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CASE REPORT:

CHORIOCARCINOMA

Mukesh Mittal*, Pankaj Kumar Nitharwal, Dinesh Kumar Sahu***, Prachi Maheshwari*****

ABSTRACT

Choriocarcinoma is an aggressive, highly vascular tumor. When it is associated with gestation, it is often considered part of the spectrum of gestational trophoblastic disease; it is then termed as gestational choriocarcinoma. While non-gestational choriocarcinoma is not associated with prior gestation and its most common site is ovary or testis. Metastasis to liver, lung, brain and vagina may occur due to vascular invasion by tumor. Initial diagnosis of Gestational trophoblastic disease is based on a multi factorial approach consisting of clinical features, serial quantitative human chorionic gonadotropin (β-hCG) titers, and imaging findings. Ultrasonography is the modality of choice for initial diagnosis of complete hydatidiform mole and can provide an invaluable means of local surveillance after treatment. Gestational trophoblastic disease after a molar pregnancy is usually diagnosed with serial β-hCG titers; imaging plays an important role in evaluation of local extent of disease and systemic surveillance. Radio-imaging is an important diagnostic tool for diagnosis of choriocarcinoma with extent of its metastasis and management of its complications like uterine and pulmonary arteriovenous fistulas. Familiarity with the pathogenesis, classification, imaging features, and treatment of these tumors can aid in radiological diagnosis and guide to appropriate management while preserving fertility.

INTRODUCTION

Choriocarcinoma is an unusual malignant tumor having an incidence of 1 in 30,000 pregnancies. Although it may occur after any pregnancy, the most important risk factor for choriocarcinoma is molar pregnancy. Spectrum of Gestational trophoblastic disease (GTD) include both malignant and benign gestational tumors which can be complete or partial hydatidiform mole, invasive mole, placental site trophoblastic tumor, epithelioid trophoblastic tumor and choriocarcinoma[1]. The later four entities are referred to as gestational trophoblastic neoplasia (GTN). These tumors are aggressive tumors and metastasise to even distant organs. GTN can result in significant morbidity and mortality if left untreated. Early diagnosis of GTN with presence or absence of metastasis is important for successful management while preserving fertility and good prognosis. Molar gestations precede 50% to 80% of cases, and 1 in 40 molar pregnancies gives rise to choriocarcinoma. Choriocarcinoma is a purely cellular lesion characterized histological by the invasion of the myometrium by abnormal, proliferating trophoblast and the absence of formed villi. Hemorrhage and necrosis are prominent features. Early vascular invasion is common, resulting in distant metastases, most frequently affecting the lungs, followed by the liver, brain, gastrointestinal tract, and kidney. Respiratory compromise may be the initial presentation followed by venous invasion and retrograde metastasis.

Choriocarcinoma is a form of malignant GTD. It arises from cytotrophoblast as well as syncytiotrophoblast without villi, and produces human chorionic gonadotropin (Beta-hCG). By vascular metastasis it can metastases to liver, lung, brain and vagina[2]. Early diagnosis by various modalities of radiological investigations is crucial for deciding the modality of treatment, its prognosis and tumor response to chemotherapy[3].

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CASE REPORT

A 27 year old P1L1woman came with the complaints of bleeding per vagina since 10 months, and lower abdominal pain since 3 weeks. Patient underwent normal vaginal delivery one year ago, after which she complaint of intermittent bleeding. On USG examination a vascular RPOC was detected for which evacuation was done. Patient underwent the evacuation for 4 times for intermittent bleeding per vagina but it persisted. Her Serum Beta hCG was raised (value – 7901 mIU/ml). On 2 D transabdominal ultrasound and 3 D Transvaginal ultrasound, uterus was bulky and a large 66 x 56 mm sized heterogenous hyperechoic mass lesion was seen in endometrial cavity with few internal cystic areas, with significant increase in vascularity on colour doppler. The lesion was invading into the myometrium causing thinning of myometrium. Bilateral ovaries were visualised normally.

On MRI, a large moderately T1W hypointense, T2 W hyperintense endocavitory lesion of size 61 x 63 x 63mm was seen. Loss of endo-myometrial junction with focal site of myometrial infiltration was identified with the depth of invasion exceeding 50% of the myometrial thickness along the anterior wall. The lesion was showing mild diffusion restriction. There was no involvement of parametrium seen.

On CECT abdomen and thorax, mild contrast enhancement was seen without any evidence of pulmonary and other soft tissue metastasis. Features were in favour of neoplastic etiology of the endometrium.

Patient underwent diagnostic and therapeutic hysteroscopy suggestive of endometrial mass. Diagnosis of choriocarcinoma was confirmed on histopathology.

DISCUSSION

Choriocarcinoma is the malignant proliferation of syncytiotrophoblast, cytotrophoblast and intermediate trophoblast with absence of chorionic villi and its direct invasion into the myometrium. It is the most aggressive type of gestation trophoblastic neoplasia (GTN) with propensity to widely metastasize. It can also be non-gestational, occurring in the cervix, ovaries, and testes or outside the reproductive tract. About 50% of gestational choriocarcinoma arise from hydatiform moles; 25% are associated with term/preterm gestation, and the remaining 25% follow abortion or tubal pregnancy. Initial diagnosis of gestational trophoblastic disease is based on a combination of history, quantitative β-hCG titer, and pelvic sonography. After that a metastatic workup with CT thorax and abdomen and MRI of pelvis is undertaken and then proved on histopathological examination[4].

CONCLUSION

Gestational choriocarcinoma is an aggressive tumor with a propensity to widely metastasize and can result in significant morbidity and mortality if left untreated. Early diagnosis of Gestational choriocarcinoma is an essential for prompt and successful management while preserving fertility. Ultrasonography is the modality of choice for initial diagnosis, management of complications such as uterine and pulmonary arteriovenous fistulas and local surveillance after treatment.

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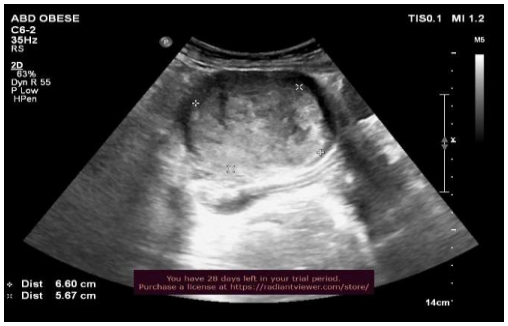

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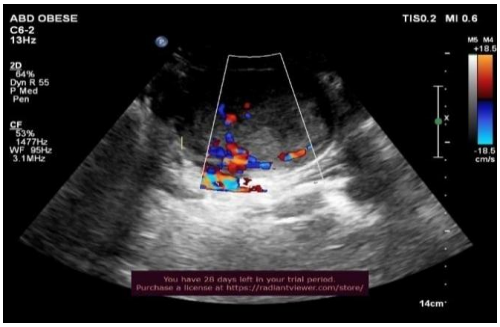

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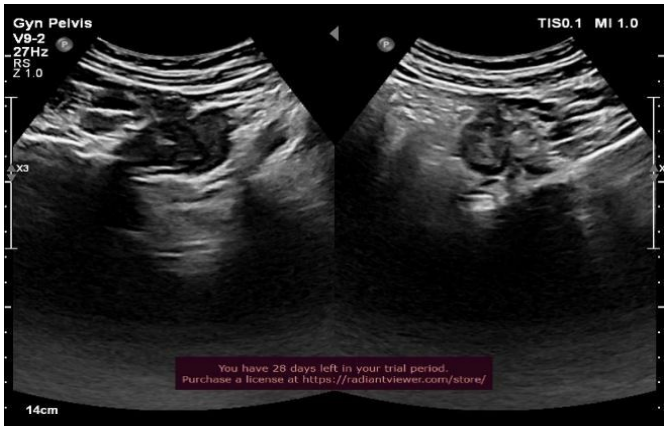

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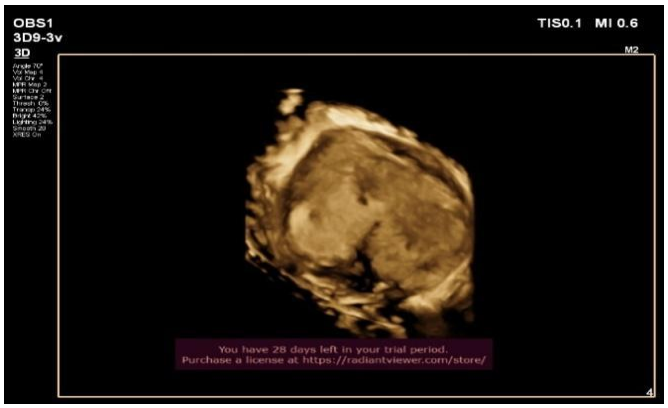
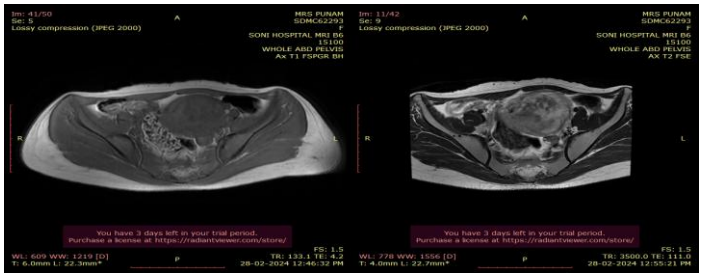
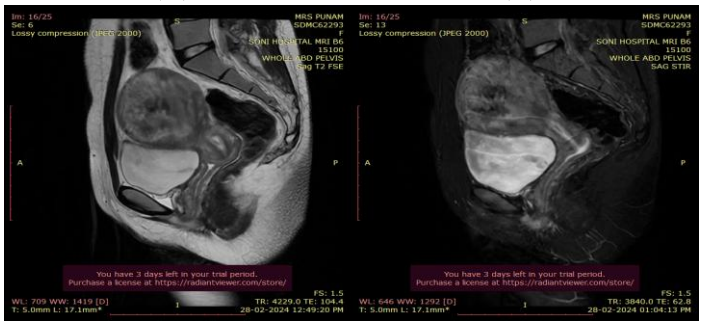


(D)

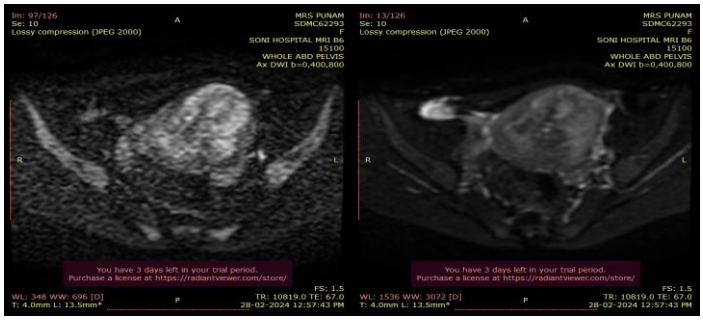
Fig 1 (A): Large heterogenous hyperechoic mass in endometrial cavity with few internal cystic areas. (B): colour doppler images showing significant increase in vascularity. (C): Bilateral ovaries seen separately. (D): 3D USG of the mass lesion showing bulky uterus with myometrial involvement.



(A) (B)



(C) (D)



(E) (F)

Fig 2. On cross sectional imaging a T1WI large hypointense (A), T2WI heterogenous hyperechoic endocavitatory mass on axial (C) and sagittal (D) with myometrial invasion, without any significant suppression on STIR (D), showing mild diffusion restriction (E,F).

CASE REPORT

CYSTIC HYGROMA

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ABSTRACT

Cystic hygroma also termed as Cystic lymphangioma is a type of lymphangioma. Cystic hygroma is a benign and infiltrative congenital malformation of the lymphatic channels with large macroscopic cystic spaces. The etiology of cystic hygroma is either developmental defect or primary due to multilocular cystic malformation of dilated lymphatic channels. Cystic hygroma is rare congenital anomaly that occurs in the usually found in the posterior triangle of the neck as a large and diffuse swelling. They usually cross the midline, reaching axillary and mediastinum. Location of Cystic hygroma corroborates the complexity and extensiveness of the lymphatic system in the cervical region in comparison to other regions of the body. The cystic hygroma can occur in cervical region (75–90%), axillary region (20%), and inguinal region and rarely at retroperitoneal and thoracic region. They usually appear as solitary lesions. They are usually infiltrative, often separating the planes of fascia and incorporating nerves, muscles, and blood vessels. They are usually painless, freely mobile, compressible, fluctuant, and trans-illuminate to light exposure. The overlying skin on the cystic hygroma is normal.

INTRODUCTION

Cystic hygroma (CH) is a congenital lymphatic malformation affecting the different parts of fetus, mostly in the fetal neck, axilla, thoracoabdominal wall and rarely mediastinal, inguinal and retroperitoneal areas. Etiology of CH is a complete or partial obstruction of the lymphatic system which prevents communication of lymphatics with the venous system and results in formation of cysts. The hygroma describes an endothelial lined mass consisting of small to medium sized lumen containing lymphatic fluid, together with a mixture of loose collagen tissue, adipose tissue and occasionally vascular tissue. The cysts may be unilocular but more often it contains multiple cysts infiltrating the surrounding structures and distorting the local anatomy[1].

An increased volume of flow in the venous system results from the lack of lymphatic drainage which leads to large veins. A complete obstruction in the lymphatic sacs prevents communication with the venous system and causes lymph fluid to accumulate and dissect into the tissues which leads to CH with septa thus create large multilocular cyst. CH without septa may result from a temporary accumulation of lymphatic fluid due to incomplete obstructions of lymphatic drainage. Increased lymphatic pressure may sometimes overcome the incomplete obstructions, thus explaining the higher spontaneous resolution rate reported with CH without septa.[2]

Fetal aneuploidy, structural malformations, hydrops fetalis and intrauterine fetal death can be associated with CH. Association of trisomies, cardiac anomalies, Turner syndrome and Noonan syndrome can occur with CH. Congenital fetal CH can be isolated lymphangiomas diagnosed in late second trimester and those which are usually associated with other malformations associated with a poor prognosis and diagnosed early in pregnancy. In the CH with septa the prognosis is considered even worse than in the CH without septa. Axillary CH was rarely reported and diagnosed often in late second trimaster[3].

CASE REPORT

A primigravida patient came for a routine antenatal scan at gestational age of 13 weeks. Previous antenatal scan of patient at 5 weeks gestational age showed no abnormality. On trans-abdominal scan findings were a single viable foetus with a well-defined anechoic cystic area with few internal septations noted around the foetal neck. It showed no flow on Colour Doppler sonography. Features of hydrops fetalis were also seen like adnominal ascites, pleural effusion, subcutaneous and scalp edema. There was no history of maternal illness or known exposure to any medication during pregnancy.

DISCUSSION

Fetal CH is a rare slow growing benign congenital developmental malformation of lymphatic system with distended fluid-filled spaces typically in the fetal neck region. Embryonic development of the lymphatic system starts at 6 weeks of gestation and later on it combines with the venous system. The inability of combination between venous system and lymphatic system results in CH. Association of CH is seen with various structural malformations including fetal aneuploidy, hydrops fetalis

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and it can lead to intrauterine fetal death. Incidence of cystic hygroma are 1 in 6000 to 1 in 16,000 live births but it is estimated to be much more than this proportion as we don't take into account the abortions (1/875)[4].

Axillary CH is rarely reported and determined often by sonographically in second trimester. Axillo-thoracic CH may be diagnosed during routine antenatal ultrasound follow-up. These are simple or multicystic structured masses, anechoic with thin septa and they may contain solid components. Usually, on Doppler examination, vascular flow isn't appreciated. Further imaging with MRI can be done for tumour extension and to determine the characteristics of the tissue. Axillary lymphangiomas account for approximately 10% of all lymphangiomas. Out of them only a few cases have been reported as diagnosed antenatally. A few cases of axillary lymphangioma causing shoulder dystocia have been diagnosed and reported after delivery. USG is the mostly commonly used diagnostic method especially for antenatal diagnosis of cervical CH. Computed tomography is not used due to harmful teratogenic effect of its ionizing radiation on mother and foetus. For further imaging MRI can be used for the anatomy and to see the extension of the tumour[5].

CONCLUSION

Cystic hygroma is a benign and infiltrative congenital malformation of the lymphatic channels. Hydrops fetalis with cystic hygroma in the second trimester has been associated with adverse outcome of foetus. Care must be taken in the fetus for early diagnosis of cystic hygroma so that counselling of patient can be done and early termination of pregnancy can be performed if the patient desires. This case is an uncommon case of Cystic Hygroma with associated features of Hydrops Fetalis. This subtype has a poor prognosis. Early antenatal diagnosis by sonography is important so that counselling of patient can be done and early termination of pregnancy can be performed if the patient desires and if patient wants to continue the pregnancy, then post delivery management based on the severity of the condition can be done.

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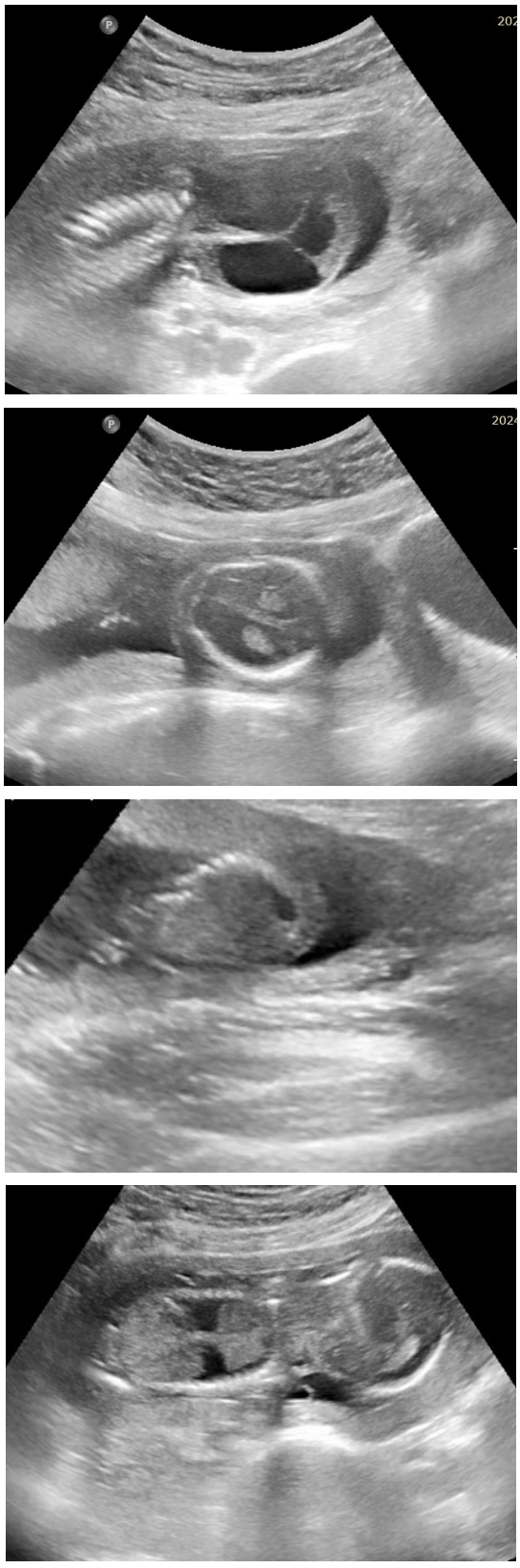

A.
B.
C.
D.

Fig 1. Anechoic cystic area around both side of the neck which is extending beyond the lateral limits of the foetus, showing internal septa (A), dilated ventricles (B), ascites (C), Pleural effusion (D).